



Summary of Core/Supplemental – Highly Recommended Recommendations: Chiari I Malformation CDEs

Start-up Resource – NINDS Chiari I Malformation CDE Recommendations

Each CDE or instrument could be classified according to the definitions below:

General Core: A data element that is required for all NINDS funded studies.

Disease Core: A data element that collects essential information applicable to any disease-specific study, including all therapeutic areas. The NINDS and its appointed working groups assign the disease “Core” classification based on the current clinical research best practices. In each case, the disease Core CDEs are a small subset of the available CDEs, where it is anticipated that investigators will need to collect the disease Core CDEs on any type of study. These are required for all disease-specific studies.

Disease Supplemental - Highly Recommended: A data element which is essential based on certain conditions or study types in clinical research studies. In most cases, these have been used and validated in the disease area. These data elements are strongly recommended for the specified disease condition, study type or design.

Disease Supplemental: A data element which is commonly collected in clinical research studies. Use depends upon the study design, protocol or type of research involved.

Disease Exploratory: A data element that requires further validation, but may fill current gaps in the CDEs and/or substitute for an existing CDE once validation is complete. Such data elements show great promise, but require further validation before they are ready for prime-time use in clinical research studies. They are reasonable to use with the understanding that it has limited validation in the target group.

<p>NIH Resources: <i>The NINDS also strongly encourages researchers to use these NIH developed materials for NINDS-sponsored research, when appropriate. Utilization of these resources will enable greater consistency for NINDS-sponsored research studies. These tools are free of charge.</i></p>	<p>NIH Toolbox Patient-Reported Outcomes Measurement Information System (PROMIS) Quality of Life in Neurological Disorders (Neuro-QOL)</p>
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Core CDEs for all NINDS Studies¹:

CDE Domain	CDE Name	CDE ID	Classification	Study Type
Demographics	Birth date	C00007	CORE	All studies
Demographics	Ethnicity USA category	C00020	CORE	All studies
Demographics	Race USA category	C00030	CORE	All studies
Demographics	Gender Type	C00035	CORE	All studies
General Health History	Medical history condition text	C00322	CORE	All studies
General Health History	Medical history condition SNOMED CT code	C00313	CORE	All studies

Core CDEs for Chiari I Malformation Studies:

Domain; Sub-Domain	Data element	CDE ID
Participant Characteristics; Demographics	Birth date	C00007
Participant Characteristics; Demographics	Gender Type	C00035
Participant Characteristics; Demographics	Ethnicity USA category	C00020
Participant Characteristics; Demographics	Race USA category	C00030
Participant Characteristics; Demographics	Medical history condition SNOMED CT code	C00313
Participant Characteristics; Demographics	Medical history condition text	C00322
Disease/Injury Related Events; Classification	Subject ID	C19247

¹ Note: Education year count C00015 is no longer a general Core CDE

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Disease/Injury Related Events; History of Disease/Injury Event	Injury date time	C05400
Assessments and Examinations; Physical/Neurological Examination	Base head pressure indicator	C22195
Assessments and Examinations; Physical/Neurological Examination	Cough cry laugh sneeze pain worse indicator	C22205
Assessments and Examinations; Imaging Diagnostics	Imaging cerebellar tonsil lowest position measurement	C21728
Assessments and Examinations; Imaging Diagnostics	Imaging cerebellar tonsil right position measurement	C21729
Assessments and Examinations; Imaging Diagnostics	Imaging cerebellar tonsil left position measurement	C21730
Outcomes and End Points; Assessing Comorbidities	Spina bifida diagnosis indicator	C22399
Outcomes and End Points; Assessing Comorbidities	Hereditary disorder connective tissue diagnosis indicator	C22402
Outcomes and End Points; Assessing Comorbidities	Cutaneous stigmata occult tethered spinal cord hair tuft indicator	C22403
Outcomes and End Points; Assessing Comorbidities	Cutaneous stigmata occult tethered spinal cord dimple indicator	C22404
Outcomes and End Points; Assessing Comorbidities	Cutaneous stigmata occult tethered spinal cord capillary hemangioma indicator	C22405
Outcomes and End Points; Assessing Comorbidities	Cutaneous stigmata occult tethered spinal cord lipoma indicator	C22406
Outcomes and End Points; Assessing Comorbidities	Cutaneous stigmata occult thread spinal cord appendage indicator	C22407

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Outcomes and End Points; Assessing Comorbidities	Cutaneous stigmata occult tethered spinal cord meningocele manque indicator	C22408
Outcomes and End Points; Assessing Comorbidities	Skin abnormality stretch mark indicator	C22409
Outcomes and End Points; Assessing Comorbidities	Skin abnormality hyperextensibility indicator	C22410
Outcomes and End Points; Assessing Comorbidities	Skin abnormality peizogenic papule indicator	C22411
Outcomes and End Points; Assessing Comorbidities	Chiari malformation family history indicator	C22415
Outcomes and End Points; Assessing Comorbidities	Injury whiplash indicator	C21887
Outcomes and End Points; Assessing Comorbidities	Traumatic Brain Injury symptom indicator	C01058
Outcomes and End Points; Assessing Comorbidities	Motor vehicle accident injury indicator	C22429
Outcomes and End Points; Assessing Comorbidities	Brain injury unspecified indicator	C22430
Outcomes and End Points; Assessing Comorbidities	Neck injury unspecified indicator	C22431
Outcomes and End Points; Assessing Comorbidities	Hydrocephalus family history untreated indicator	C22447
Outcomes and End Points; Assessing Comorbidities	Hydrocephalus family history treated indicator	C22448
Outcomes and End Points; Assessing Comorbidities	Hydrocephalus family history slit ventricle indicator	C22449

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Outcomes and End Points; Assessing Comorbidities	Hydrocephalus family history aqueductal stenosis indicator	C22450
Outcomes and End Points; Assessing Comorbidities	Hydrocephalus family history ventricle variant indicator	C22451
Outcomes and End Points; Assessing Comorbidities	Klippel Feil Anomaly diagnosis indicator	C22452
Outcomes and End Points; Assessing Comorbidities	Gravida maternal value	C11065
Outcomes and End Points; Assessing Comorbidities	Pregnancy miscarriage medical termination still birth count	C22456
Outcomes and End Points; Assessing Comorbidities	Parity maternal value	C11063
Outcomes and End Points; Assessing Comorbidities	Medical history Menieres disease indicator	C22459
Outcomes and End Points; Assessing Comorbidities	Medical history cervical radiculopathy disc disease indicator	C22465
Outcomes and End Points; Assessing Comorbidities	Medical history infectious disease central nervous system implication indicator	C22466
Outcomes and End Points; Assessing Comorbidities	Medical history sarcoidosis condition indicator	C22467
Outcomes and End Points; Assessing Comorbidities	Medical history tuberculosis condition indicator	C22469
Outcomes and End Points; Assessing Comorbidities	Medical history Lyme disease indicator	C21870
Outcomes and End Points; Assessing Comorbidities	Medical history cerebrospinal fluid leak intracranial hypotension indicator	C22474

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Outcomes and End Points; Assessing Comorbidities	Medical history spinal stenosis indicator	C22475
Outcomes and End Points; Assessing Comorbidities	Spinal cord tethered none indicator	C22476
Outcomes and End Points; Assessing Comorbidities	Spinal cord tethered lipoma indicator	C22479
Outcomes and End Points; Assessing Comorbidities	Spinal cord tethered myelomeningocele indicator	C22480
Outcomes and End Points; Assessing Comorbidities	Spinal cord tethered thick filum indicator	C22481
Outcomes and End Points; Assessing Comorbidities	Medical history depression since last visit indicator	C19877
Outcomes and End Points; Assessing Comorbidities	Psychiatric anxiety indicator	C19831
Outcomes and End Points; Assessing Comorbidities	Attention deficit hyperactivity disorder indicator	C22478

General Core Instrument for all FDA-regulated Chiari I malformation clinical trials:

Investigators should review the FDA's ["Guidance for Industry: Suicidal Ideation and Behavior: Prospective Assessment of Occurrence in Clinical Trials"](#) for the most up-to-date information about suicidal ideation and behavior. One scale that FDA suggests is the Columbia Suicide Severity Rating Scale (C-SSRS) (available at [Columbia Suicide Severity Rating Scale Website](#)).

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Supplemental – Highly Recommended CDEs for Chiari I Malformation Studies:

Domain; Sub-Domain	Data element	CDE ID
Participant History and Family History; General Health History	Imaging diagnosis stroke transient ischemic attack indicator	C13758
Participant History and Family History; General Health History	Medical history visual impairment indicator	C21872
Participant History and Family History; General Health History	Medical history hearing deficit indicator	C21873
Participant History and Family History; General Health History	Medical history paraplegia quadriplegia indicator	C21874
Participant History and Family History; General Health History	Surgical history cataract removal indicator	C21882
Participant History and Family History; General Health History	Pain lower back indicator	C21884
Participant History and Family History; General Health History	Pain general leg indicator	C21885
Participant History and Family History; General Health History	Injury head indicator	C21886
Participant History and Family History; General Health History	Injury whiplash indicator	C21887
Participant History and Family History; General Health History	Injury fall head neck indicator	C21888

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Participant History and Family History; General Health History	Echocardiogram mitral valve prolapse indicator	C12875
Participant History and Family History; General Health History	Medical history aortic root enlargement indicator	C22433
Participant History and Family History; General Health History	Beighton Score - Total score	C21689
Participant History and Family History; General Health History	Hereditary disorder connective tissue diagnosis indicator	C22402
Participant History and Family History; General Health History	Hereditary disorder type	C22776
Participant History and Family History; General Health History	Hereditary connective tissue disorder diagnosis type	C22775
Participant History and Family History; General Health History	Chiari malformation screening hypermobility family indicator	C21761
Participant History and Family History; General Health History	Chiari malformation screening echocardiogram mitral valve prolapse aortic aneurysm diagnosis indicator	C21766
Participant History and Family History; General Health History	Chiari malformation screening orthopedic joint hip dislocation unusual pain screen indicator	C21767
Disease/Injury Related Events; Classification	Chiari I malformation headache pressure daily occurrence indicator	C21690

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Disease/Injury Related Events; Classification	Chiari I malformation neck pain stiff radiate shoulder symptom indicator	C21691
Disease/Injury Related Events; Classification	Chiari I malformation prickle tingle numb arm leg frequent occurrence symptom indicator	C21692
Disease/Injury Related Events; Classification	Chiari I malformation position change dizzy symptom indicator	C21693
Disease/Injury Related Events; Classification	Chiari I malformation unsteady stand walk symptom indicator	C21694
Disease/Injury Related Events; Classification	Chiari I malformation headache pressure eye ear light sensitive symptom indicator	C21695
Disease/Injury Related Events; Classification	Chiari I malformation headache strain activity worsen symptom indicator	C21696
Disease/Injury Related Events; Classification	Chiari I malformation chronic fatigue short-term memory loss irritable symptom indicator	C21697
Disease/Injury Related Events; Classification	Chiari I malformation generalize weakness stiff arm leg symptom indicator	C21698
Disease/Injury Related Events; Classification	Chiari I malformation shortness breath blur vision ring ear urinary urgency heart palpitation symptom indicator	C21699
Disease/Injury Related Events; History of Disease/Injury Event	Headache primary anatomic site	C21778

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Disease/Injury Related Events; History of Disease/Injury Event	Headache primary laterality anatomic site	C21780
Disease/Injury Related Events; History of Disease/Injury Event	Headache primary laterality anatomic site score	C21782
Disease/Injury Related Events; History of Disease/Injury Event	Headache frequency type	C21790
Disease/Injury Related Events; History of Disease/Injury Event	Headache frequency score	C21791
Disease/Injury Related Events; History of Disease/Injury Event	Headache duration type	C21837
Disease/Injury Related Events; History of Disease/Injury Event	Headache duration score	C21838
Disease/Injury Related Events; History of Disease/Injury Event	Headache aggravating pain factor type	C21842
Disease/Injury Related Events; History of Disease/Injury Event	Headache aggravating pain factor score	C21843
Disease/Injury Related Events; History of Disease/Injury Event	Headache assessment total score	C21844
Assessments and Examinations; Physical/Neurological Examination	Behind eye pain radiate indicator	C22198
Assessments and Examinations; Physical/Neurological Examination	Neck shoulder pain radiate indicator	C22200
Assessments and Examinations; Physical/Neurological Examination	Stiffness pain general indicator	C22208
Assessments and Examinations; Physical/Neurological Examination	Chiari I malformation position change dizzy symptom indicator	C21693

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Assessments and Examinations; Physical/Neurological Examination	Swallow difficulty indicator	C22722
Assessments and Examinations; Physical/Neurological Examination	Respiration stop during sleep indicator	C21577
Assessments and Examinations; Physical/Neurological Examination	Loss of consciousness indicator	C01052
Assessments and Examinations; Physical/Neurological Examination	Chiari I malformation prickle tingle numb arm leg frequent occurrence symptom indicator	C21692
Assessments and Examinations; Physical/Neurological Examination	Pain touch sensitivity increase indicator	C22729
Assessments and Examinations; Physical/Neurological Examination	Pain sensitivity decrease indicator	C22730
Assessments and Examinations; Physical/Neurological Examination	Sensation loss extremities indicator	C22731
Assessments and Examinations; Physical/Neurological Examination	Burning pain abnormal extremity indicator	C22732
Assessments and Examinations; Physical/Neurological Examination	Extremity specific area pain decreased sensation indicator	C22733
Assessments and Examinations; Physical/Neurological Examination	Extremity weakness indicator	C22736
Assessments and Examinations; Physical/Neurological Examination	Muscle tone loss indicator	C22737
Assessments and Examinations; Physical/Neurological Examination	Joint hypermobility indicator	C22764
Assessments and Examinations; Physical/Neurological Examination	Beighton Score - Motion thumb forearm assessment score	C21684

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Assessments and Examinations; Physical/Neurological Examination	Beighton Score - Motion finger bend back score	C21685
Assessments and Examinations; Physical/Neurological Examination	Beighton Score - Motion elbow hyperextend score	C21686
Assessments and Examinations; Physical/Neurological Examination	Beighton Score - Motion knee hyperextend score	C21687
Assessments and Examinations; Physical/Neurological Examination	Beighton Score - Motion palm flat leg straight score	C21688
Assessments and Examinations; Physical/Neurological Examination	Beighton Score - Total score	C21689
Assessments and Examinations; General and Motor	Reflex autonomic reflex valsalva maneuver status	C22365
Assessments and Examinations; Imaging Diagnostics	Imaging syrinx present indicator	C21732
Treatment/Intervention Data; Drugs	Medication prior name	C18035
Treatment/Intervention Data; Drugs	Medication prior or concomitant dose	C02006
Treatment/Intervention Data; Drugs	Medication prior or concomitant dose frequency	C02011

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Treatment/Intervention Data; Drugs	Medication prior or concomitant start date and time	C02016
Treatment/Intervention Data; Drugs	Medication prior or concomitant end date and time	C02008
Treatment/Intervention Data; Therapies	Imaging post surgical procedure type	C22370
Treatment/Intervention Data; Therapies	Imaging pre surgical procedure type	C22373
Treatment/Intervention Data; Therapies	Tonsil lowest position measurement	C22376
Treatment/Intervention Data; Therapies	Tonsil right cerebellar measurement	C22377
Treatment/Intervention Data; Therapies	Tonsil left cerebellar measurement	C22393
Treatment/Intervention Data; Therapies	Syrinx width measurement	C22518

Supplemental – Highly Recommended Instruments:

1. Nurick (Ambulatory Function) Grade
2. ASIA Motor Scale
3. Headache Disability Index
4. Meaningful Improvement

For the complete list of NINDS CDE recommendations for Chiari I Malformation, please see the [NINDS CDE website](#).